Atypical *Truncus Arteriosus* Operated at 28 Years of Age: Importance of Differential Diagnosis

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This is the case of 28 year-old adult with suspected congenital heart disease since birth, not treated in childhood at the his family's choice. At 27 years old, he was diagnosed with pulmonary atresia with ventricular septal defect and systemic-pulmonary collaterals, where surgery was contraindicated. A new review in our department showed that it was an atypical form of *truncus arteriosus*. The fact that a common arterial trunk with left-right shunt was viewed by echocardiography was a crucial fact for the indication of new catheterization, opening the prospect of surgical correction.

Currently, the patient is well, with 7 years of postoperative outcome.

Introduction

The *truncus arteriosus* or common arterial trunk, is a form of cyanotic heart disease where only one artery arises from the heart, being responsible for the systemic, pulmonary and coronary circulation[1]. It occurs in 1.5% of cases of congenital heart defects in newborns, presenting variations in their presentation as to the origin of the pulmonary trunk, which in 1949 generated an initial classification by Collett and Edwards[2], followed by Van Praagh[3]. Rare forms that do not fall into these classifications may occur.

Although several cases of diagnosis in foetus[4,5] life have been described, the diagnosis is made in the neonatal period or in childhood. Surgical correction is done in childhood and rarely in adult life[6,7].

Case Report

Male adult, 28 years old, with suspected congenital heart disease from birth not investigated due to refusal of parents, was acyanotic and satisfactory developed up to 10 years of age, when an episode of pneumonia led the patient to a referral center. Cardiac catheterization was performed, but the result has never been known by the family due to abandonment of treatment.

At 27, the patient began to experience nocturnal dyspnea and was referred to a reference center, where he underwent echocardiography, who diagnosed pulmonary atresia with wide ventricular septal defect (VSD), moderate aortic insufficiency, important ectasia of the aortic root. A new catheterization confirmed the echocardiographic findings, also demonstrating the presence of systemic-pulmonary collateral arteries. At that time, surgery was contraindicated.

After five months, the patient came to our hospital for a second opinion. The echocardiogram suggested the diagnosis of atypical form of *truncus arteriosus*, due to a large vessel emerging before the ascending aorta, giving rise to the pulmonary trunk, which did not fit the classically described types[1,2] (Figures 1A and 1B).

The color mapping showed accelerated flow from the truncus to the pulmonary trunk, suggesting low pulmonary vascular resistance. The truncal valve was tricuspid, slightly insufficient and the coronary arteries emerged in separate ostia with no abnormalities.

Based on echocardiographic findings, a third cardiac catheterization was performed, which confirmed the diagnosis of *truncus arteriosus* without specifying type, associated with the artery to the middle lobe of right lung originating from the descending aorta with stenosis at the origin, severe hypertension in the right chambers (100/50/70 mmHg) and normal pressure in the aorta (120/55/80 mmHg). After the catheterization, the patient was discharged as the surgical team in charge of his case concluded that there was no indication for surgery.

After two months, a third opinion was requested by the physician from the original center that referred the patient to our group. On this occasion, the patient was in good general condition, acyanotic, cardiac auscultation with a continuous murmur ++/4 throughout the precordium accompanied by fremitus. The electrocardiogram showed sinus rhythm and diffuse changes in ventricular repolarization. The radiographic image showed an enlarged cardiac silhouette +++/4 at the expense of the right ventricle with congested pulmonary vascular segment, especially in the perihilar region and right lung base (Figure 1C). An magnetic resonance imaging confirmed the presence of a pulmonary trunk with sinus origin (below the sinotubular junction) and stenosis prior to the bifurcation (Figure 1D), ruling out the possibility of this being a collateral vessel emerging from a coronary artery.

Keywords

Heart defects, congenital/diagnosis; pulmonary atresia; heart septal defects, ventricular; *truncus arteriosus*.

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Case Report

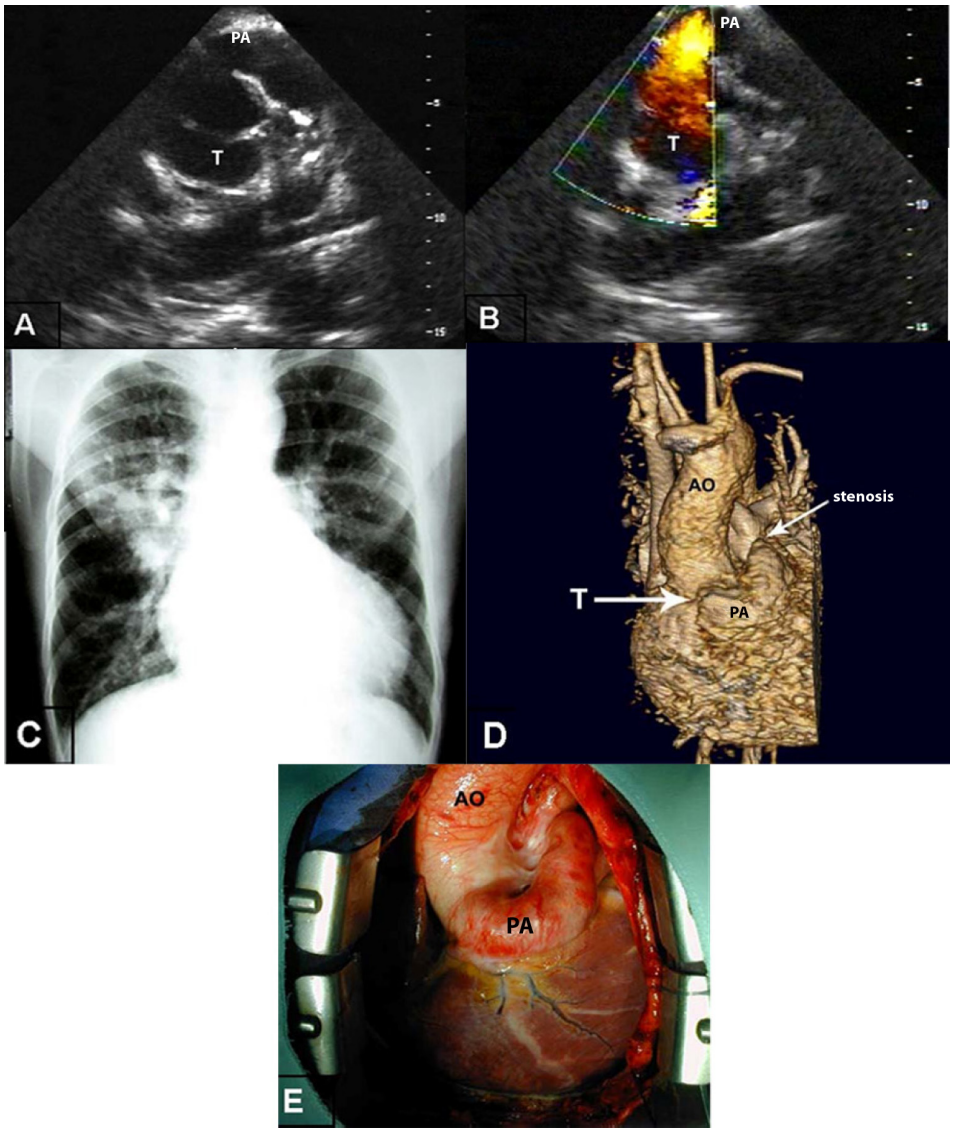

Figure 1 - *A) Echocardiography showing pulmonary artery emerging from the anterior portion of the common arterial trunk; B) Flow in red indicating the left-right direction of the flow, from the truncus into the pulmonary artery; C) Radiographic image showing an enlarged cardiac silhouette and congested pulmonary vascular segment; D) Magnetic resonance imaging showing large pulmonary artery emerging from the anterior portion of the common arterial trunk. The arrow indicates the site of pulmonary stenosis before the bifurcation; E) Same aspect observed during surgery. T - Truncus arteriosus; AP=PA - pulmonary artery, AO - aorta.*

Given these findings, we decided for total correction. The surgery confirmed the sinus origin of the pulmonary trunk near the right coronary artery with stenosis before bifurcation, and an intense network of collateral arteries (Figure 1E). The surgery was uneventful. The right ventricle outflow tract (RV) was reconstructed with bovine pericardium, implantation of bioprosthesis No. 27 in the pulmonary position, closure of VSD and ligation of major descending aorta to the left lung (Figures 2A, 2B, 2C and 2D).

The patient evolved with low output and heart failure due to biventricular dysfunction, requiring vasoactive drugs for long periods with gradual recovery of functions. He was discharged 50 days after surgery.

In seven years of late postoperative follow-up, although showing good clinical outcome, cardiac catheterization was indicated in order to study the pulmonary pressures and embolize the collateral artery to the right middle lobe. Right ventricular pressures measured 40/0/5 mmHg. As this collateral artery was stenotic at origin and because there were no pulmonary artery branches to this lobe, we opted for expectant management (Figures 2E, 2F and 2G).

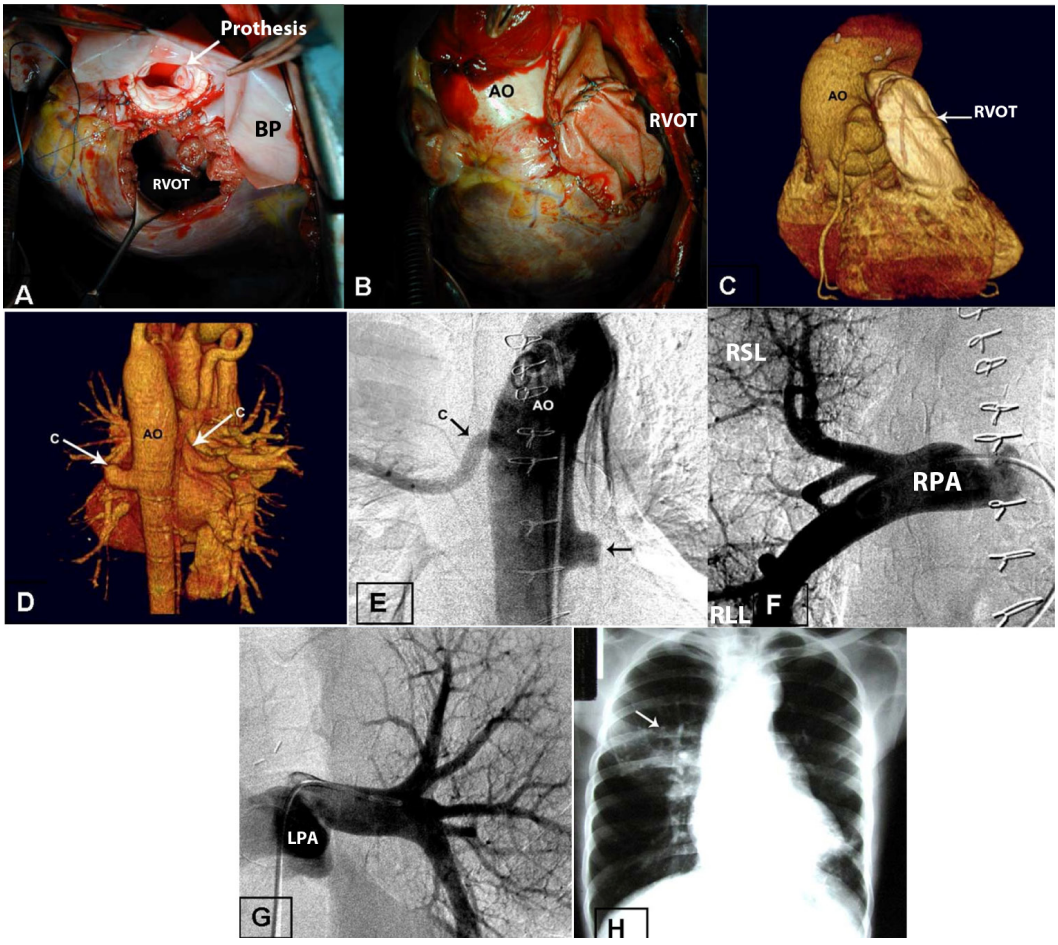

Figure 2 - *A) Heart during surgery with right ventricle open, visible bioprosthesis and bovine pericardium which was used in the reconstruction of right ventricle outflow tract; B) Final aspect of the right ventricle outflow tract reconstruction surgery, also shown in C) Computed angiography of the heart; D) Descending aorta in posterior view with collateral artery to the left lung surgically closed and long collateral artery to the right middle lobe. E) Cardiac catheterization after seven years of surgery with descending aorta in anterior view with long collateral artery to the middle lobe and collateral artery to left lung surgically closed; F) Right pulmonary artery perfusing the right lung with the exception of the middle lobe, G) Left pulmonary artery; H) Radiographic image showing significant decrease in heart size and pulmonary congestion. The arrow points to the right pulmonary hilum that remains supplied by the collateral artery. RVOT - right ventricle outflow tract; BP - bovine pericardium; AO - aorta, C - systemic-pulmonary collateral artery; RPA - right pulmonary artery; RSL - right upper lobe; RLL - right lower lobe; LPA - left pulmonary artery.*

Currently, the patient is asymptomatic, making use of amiodarone to control ventricular arrhythmia, significant decrease of cardiac silhouette and pulmonary congestion at RX (Figure 2H). The current echocardiogram shows pulmonary bioprosthesis with discrete gradient of 16 mmHg, mild aortic regurgitation with recovery of biventricular function.

Discussion

This case demonstrates the importance of specialized echocardiography for adults with congenital heart disease. The first diagnosis of *truncus arteriosus* in the life of this patient, promoted by our hospital, prompted the hospital's medical staff to deepen invasive workups, even with a history of two previous cardiac catheterizations with a diagnosis of pulmonary atresia with VSD.

It is also important to emphasize the correct evaluation of data from the third cardiac catheterization, which revealed high pressures in the right chambers, logically due to transmission of systemic pressures of the common arterial trunk to both ventricles and also due to the presence of large VSD. Another important surgical indication data was that the pulmonary trunk presented severe stenosis at the origin, which protected the lung until adulthood. The presence of systemic-pulmonary collateral arteries originating from the descending aorta and viewed in previous examinations must have been one of the reasons that led to the diagnosis of pulmonary atresia with VSD. Knowing that it is a rare association of *truncus* with systemic-pulmonary collateral arteries, the most likely hypothesis to justify such an occurrence is that they have been acquired over the patient's life, 28 years old and not operated.

The good outcome was favored by the peculiar arrangement of the sinus origin of the pulmonary orifice, which, for presenting a smaller distance between the pulmonary trunk and right ventricle outflow tract, made easier the reconstruction of this region.

The authors emphasize the difficulties of a differential diagnosis in this complex case of atypical truncus.

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Potential Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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Study Association

This study is not associated with any post-graduation program.

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